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CYLINDROMA ENDOTHELIOIDES OF THE DURA MATER CAUSING LOCALIZING SYMPTOMS AND EARLY MUSCULAR ATROPHY.

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Spreading paralysis on the right side, commencing in the shoulder; Jacksonian epilepsy; absence of all the classical brain tumor symptoms; early amyotrophy without spinal lesions; change of electric reaction; operation; death.—(Dr. Carson.) P. McH., aged forty-six years, born in Ireland, resident of Kansas, occupation horse-trainer, gives the following history:

General health always good with the exception of chills and fever at various times six or seven years back. Always led an active life, and excelled in athletic sports. A little more than a year before the beginning of the present trouble he was thrown from a buggy and dragged about thirty yards. Was not conscious at the time of an injury.

One day, about six months before presenting himself for treatment, he noticed a weakness in the right shoulder, when, with some companions, he attempted to vault a fence. On repeated examinations he found that, while the shoulder and arm were decidedly weak, the elbow-joint and wrist were still performing their functions, and the grip of the hand and the movement of the fingers were not impaired. One month later the arm became so weak and unsteady, and a few weeks later the grip of the hand so unreliable, that he was compelled to discontinue shaving himself. The whole arm and hand felt at times numb (sleepy), and the arm seemed smaller. At this time he sought medical advice from a local physician. Treatment (chiefly bromides and galvanism) was of no avail, and was, therefore, soon abandoned. For several months the impairment of power and the "numbness" continued to increase.

Electrical treatment was resumed by his physician, the faradic current being applied to the affected member. This application was immediately followed by a spasm commencing in the index and middle fingers, which were flexed upon the palm, the thumb being thrown into the palm; then the spasm extended to the wrist, flexing it; then to the arm, flexing it and drawing it near to the chest. All of the flexed muscles of the fingers, hand, forearm, and arm to the shoulder were involved. The spasm was unattended with loss of consciousness, lasted about two minutes, and resisted all attempts at reduction. Such attempts caused much pain and had to be discontinued. The arm remained absolutely useless for at least an hour afterward, when it gradually resumed its former condition.

Again, during the afternoon of the same day, he had a second spasm, a counterpart of that in the morning. During the time that elapsed from the first attack to his entrance into the hospital, about three months,

he had at varying intervals nine spasms of different intensity, but in all essentials like the first one. Some of them, he thought, had been shortened by forcibly extending the fingers, their starting-point, when the first indication of return was felt. Ten days before entering the hospital he had the last attack. This was more severe than any of the others, extending from the arm into the face, closing the eye, twisting the leg as it had done the arm, and lasting about ten minutes. There had been *no* headache, vertigo, nausea, pain or suffering of any kind, nor impairment of sight, nothing to indicate that his general health was failing.

Status Præsens. At the time of entering the hospital examination revealed the following conditions: Appearance robust, body well nourished; all the faculties of the mind intact; patient is intelligent, far above the average of his class; no external evidence of injury; the left side of the head slightly larger than the right, and somewhat hypersensitive a little below and anteriorly to the left parietal eminence. The right arm completely paralyzed and the right leg paretic. Measurement shows the right arm ten and one-half and the left twelve inches in circumference. The right forearm one half inch less than the left. The right thigh is one inch and the right leg one-quarter inch less than the left. There is slight paresis of the face on the right side. All the tendon-reflexes on the right are exaggerated and foot-clonus well marked; slight spasticity in triceps of right arm; sensation and muscle sense not at all impaired. Patient is left-handed.

Examination of the eyes made by Drs. Pollack and Wolfner, of this city, independently of each other, did not reveal any abnormalities except an engorgement of the retinal veins. Accommodation and acuity of vision were perfect. There was no choked disk. The diagnosis was: Tumor of the brain, left hemisphere, involving principally the finger, wrist, and arm-centres. Since the trouble was evidently progressive, an operation was decided upon. The absence of headache, vertigo, nausea, and choked disk could not invalidate the diagnosis, which was rendered very probable by the characteristic spreading of the paralysis and the typical Jacksonian epilepsy.

The usual preparation for aseptic operation having been made, and, after chloroforming the patient, the flap having been raised in the customary manner, a button of bone was removed with an inch-trephine from the marked point, a little behind and below the middle of a line corresponding to the Rolandic fissure. Before the bone was entirely cut through there was profuse hemorrhage, which at first was supposed to come from the dilated vessels of the diploë. Another disk was removed anteriorly and a little above the former, and the two openings thus made were united by removing the intervening bridge of bone with a chisel. The hemorrhage from the entire circumference of the cranial opening was alarming, and, in order to control it, the opening had to be packed with gauze while the rongeur forceps were being used to enlarge the operative field. The copious bleeding was subsequently found to be caused by the immense size of some branches of the posterior middle meningeal vein, which, through excessive dilatation and subsequent corrosive (osteolytic) action, had converted the grooves in the skull into deep and broad channels, and it was not until the bone, including the large vessels, had been removed that the hemorrhage could be controlled.

Upon the removal of the bone the dura bulged into the opening and

presented a dark, congested appearance, being covered with many and large gorged vessels. There was neither to be seen nor felt the least indication of pulsation. Upon attempting to raise the dura it was found to be adherent to a growth beneath, which was now plainly visible and, while it was soft and friable, was easily lifted from its bed between the dura and the apparently healthy brain. The dura, that part of it involved in the tumor, was cut out and the cavity left loosely packed with gauze, the flap replaced, and held by a fine silk suture, the wound dressed, and the patient returned to bed in a bad condition on account of shock and the considerable loss of blood.

During the course of the operation injections of whiskey, aqua ammonia, and ether, had been administered by one of my assistants to prevent collapse.

The tumor, at the time of removal, was three and one-half inches long, three inches wide, one inch thick, and weighed fourteen ounces.

Patient seemed to be getting along satisfactorily during the first three days after the operation. There was dulness of sensation on the right side, which, however, seemed to have completely disappeared on the third day. The paralysis of the arm also seemed to improve, when, on the fourth day, the patient showed a brown, dry tongue, and was slightly delirious. Abscesses had formed at the sites of the hypodermatic injections on the right arm. They were opened, and the cavities cleaned with peroxide of hydrogen. After this the case progressed favorably until the twelfth day, when fever and a grave delirium, bordering at times on maniacal wildness, set in.

On the same day he had a spasm which involved the whole right side and left him unconscious. He never rallied from this, and died seventeen days after the operation. The temperature never exceeded 101.2°.

Remarks by Dr. Bremer.

Autopsy. The wound is united firmly throughout. The scalp appears sunken at the site of the operation. Upon the removal of the calvarium the dura mater is found strongly adherent to the bony edge of the trephine opening. The scalp is united to the underlying brain-substance (the dura having been removed at this place). No sign of a suppurative process. The dural vessels posteriorly to the operative field are congested and tortuous; the same is true of the corresponding portion of the pia. The margin of the dura is considerably thickened, and, as already stated, adheres firmly to the adjacent cerebral matter.

The brain surface presents an irregular, shallow defect in the nature of an erosion (Fig. 1) nearly circular in form, about two and one-half inches in diameter. It is of a pale-reddish color, and looks roughened, shreddy, and lacerated.

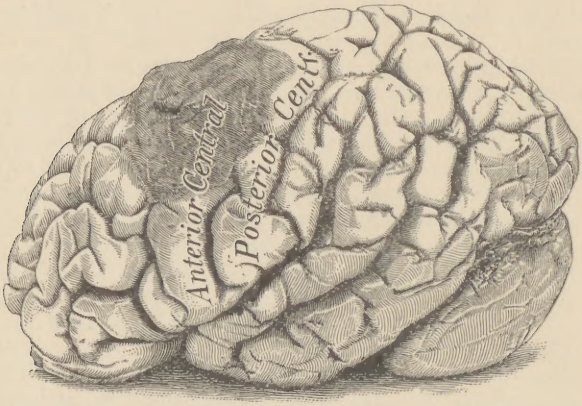
Toward the longitudinal sinus it is with greater difficulty than is usual under normal conditions that the dura mater can be separated from the subjacent tissue; a white mass about the size and shape of a split butter-bean is observed growing from the dura mater anteriorly to the defect. (This formation is recognized as being a remnant of the tumor which had escaped removal at the operation.)

There is a clot, not adherent, extending between the lesion and the longitudinal sinus, reaching backward over the superior parietal lobule for about two inches. The right hemisphere and the under surface of

the brain do not offer any abnormalities, except that the basilar artery is greatly enlarged.

The main area involved by the erosion is the greater portion of the upper two-thirds of the ascending frontal convolution and the foot of the first frontal; superiorly it commences about one-half inch from the longitudinal fissure, and extends downward to within two inches of the fissure of Sylvius. It involves the entire breadth of the ascending frontal and encroaches anteriorly on the first and second frontal convolutions, and posteriorly on the posterior ascending, which it displaces backward.

FIG. 1.



From a photograph of the brain hardened in Müller's fluid, showing the extent of the tumor. To make it appear plainer, the edges have been marked. It can be seen that the posterior central is dislocated backward. The tumor seems to reach up to the great longitudinal fissure, which, in reality, is not the case. The space between the upper margin of the tumor and the fissure is invisible, owing to the profile view.

On making a frontal incision at the site of the lesion the underlying structure on the cut surface appears reddish-brown for about an inch or an inch-and-a-half downward, shading gradually into the apparently white normal substance beneath.

The opinion prevailed at the time of the operation that the tumor had been radically removed, it being impossible to feel any remnants as far as the finger could reach with safety. Moreover, the brain seemed to be, aside from an easily comprehensible paleness and a flattened appearance of the convolutions, absolutely healthy. It was hoped, therefore, that in spite of the size of the tumor, the brain at the site of the depression would gradually resume its normal relations and functions.

This hope was not realized. Before the operation had been completed the cavity left by the removal of the tumor had been filled out by the underlying previously-compressed brain-mass, a process which, owing to its rapidity, could not very well have been accomplished without some hemorrhage into the substance of the brain.[1] Many hemorrhagic foci

[1] It seems that even the most careful handling of the healthy, or apparently healthy, brain, such as cautious palpation, is invariably attended with hemorrhage, especially into the cortical substance, owing to the extreme thinness and vulnerability of the cerebral capillaries. This I

of varying sizes were indeed found on closer examination in addition to the other changes to be described later on.

But, above all, the unremoved portion of the tumor on the visceral side of the dura would have precluded any possibility of a permanent cure, the tumor-mass gradually flattening out toward the healthy portion of the dura, and, after the manner of malignant growths, imperceptibly merging into the healthy tissue. Anteriorly to the opening in the dura the remaining mass was largest. Its thickness did not, however, taper off smoothly and evenly toward the periphery, but there was a terrace-like appearance, recalling the successive formation of the strata of ice seen around water-pumps in winter. The tumor, therefore, gave the impression of a miniature mountain planted with a broad base on the visceral layer of the dura mater.

Macro- and microscopic examination. The organs examined were the lungs, part of liver, kidneys, spinal cord, and brain. The following is the finding:

The lungs are gorged with blood. Second stage of croupous pneumonia. Liver much swollen, distinct lesions not demonstrable. Kidneys very much enlarged, cortical layer pale and thickened, medullary gorged with blood. Epithelium in many places degenerated and shed. Casts in all parts of the organ; some of them pigmented. Many hemorrhagic foci.

Spinal cord. There is macroscopically no demonstrable lesion of the cord, except a defect around the central canal, which, however, is looked upon as an artefact, due to maceration in the preserving fluid and possible rough handling during removal. Microscopically, however, this region shows a considerably greater amount of round cells around the canal than is normal, pointing to an irritating process. This extends down to the lumbar enlargement.

Very careful investigation is made as to the presence of a descending degeneration of the cortico-muscular tracts. The cervical enlargement especially is scrutinized owing to the striking atrophy of the right arm. Specimens prepared after Weigert do not show the slightest trace of any degenerative processes of the tracts mentioned nor of any nerve fibres in other fields of the section.

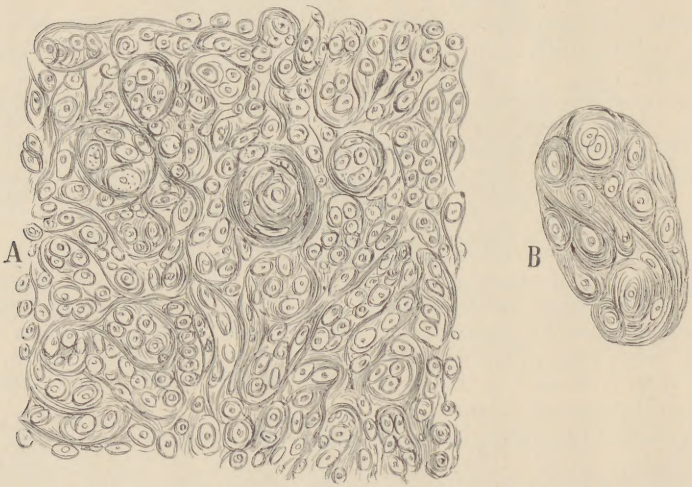
The large ganglionic cells of the anterior horns are on the whole intact, especially the peripheral ones. Some, however, of those lying more toward the centre of the horn show signs of degeneration (loss of processes and impairment of tinctorial capacity), not extensive enough, however, to account for the pronounced atrophy existing in the arm. At any rate, there is no difference in the extent of degeneration between the left and right anterior horns, which would be expected to be the case if the atrophy had been dependent on an anatomic change of the ganglionic cells. Besides, wasting of a small number of these cells nearing the age of fifty is, I think, physiologic.

was able to demonstrate in a specimen of apparently healthy cortex excised at the "wrist-centre" in a case of cortical epilepsy. Although great care had been taken by the operator, Dr. Prewitt, of this city, to avoid any undue pressure, innumerable minute capillary hemorrhages could be demonstrated microscopically throughout the excised piece of normal cortex and, to a less extent, the underlying medullary portions.

Microscopic description of the tumor. The tumor is a cylindroma endothelioides. The term cylindroma comprises a variety of tumors, and, reading up on this subject in the text-books, Ziegler's *Pathological Anatomy*, for instance, one does not get a very clear idea of the matter. By it are meant several kinds of growths having in common cylinder-like bodies of a hyaline or gelatinous nature. Most of them belong to the class of the sarcomata. So does our tumor. I have called it cylindroma endothelioides, because the tumor in the main mass and fully-matured state is an endothelioma containing the cylindroid masses mentioned. It might, in the present unsettled state of nomenclature, also be called endothelioma cylindroides or sarcomatodes, or, as we shall see directly, angio-sarcoma cylindroides.

The tumor is, indeed, somewhat of a pathologic mine of wealth. It represents an astonishing variety of the phases of sarcoma development.

FIG. 2.



Müller's fluid carmine.

A. Typical endothelioma from fully-developed portion of the tumor. (HARTNACK, 3, vi.)

B. The same with higher amplification. (HARTNACK, 3, viii.)

While, as stated before, the main mass is clearly made up of flattened or elongated cells which, after the fashion of epithelial growths, press upon each other, showing sometimes the well-known onion-like arrangement so characteristic of the epitheliomata, even the "pearl," nothing of this sort is seen in other parts of the tumor.

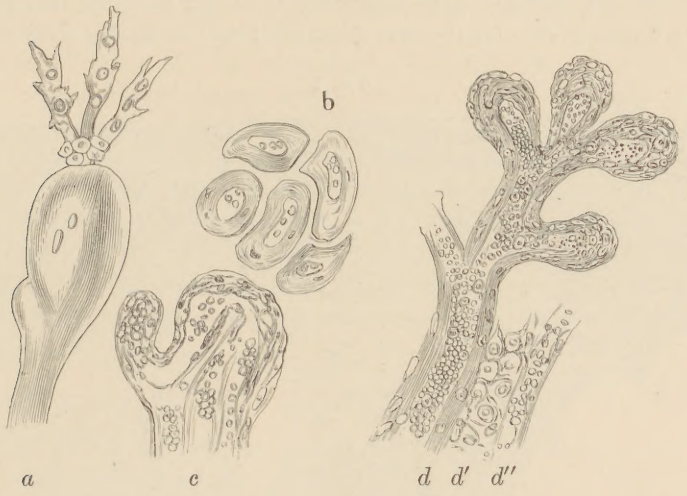
Nearly all the varieties of sarcomata may be encountered in or near the tumor mass: the large oval-celled, net-celled, myxo- and angio-sarcoma.

The origin of the cylinder-like hyaline mass is a still debated question among pathologists. Some, I believe the majority of authors on the subject, have claimed that such originate in vessels undergoing a hyaline change. There can be no question about this in our case. The vessels are plainly seen in the process of the change. This plainness is

probably due to the fact that the tumor is of recent growth. In another tumor of the same kind which I had an opportunity of examining, and which was of several years' growth, this connection between the hyaline masses and the vessels and the tumor could not be demonstrated.

In our tumor there is not only a number of vessels undergoing this process and still showing blood corpuscles in their lumen, but the general aspect of the hyaline masses at once suggests their origin.

FIG. 3.



Isolated vessels of the tumor.

a. A vascular bud in complete hyaline degeneration; at the top some isolated net cells of the tumor proper. *b*. Cross-section of a bunch of vessels with partially hyaline walls. *c*. Cactus form of growing vessels. *d*. The same with typical buds. To the right of the vessel are cells closely packed (*d′*), and a small capillary (*d″*). Small granular hæmoglobin-containing bodies are seen in the lumen of the buds on the right.

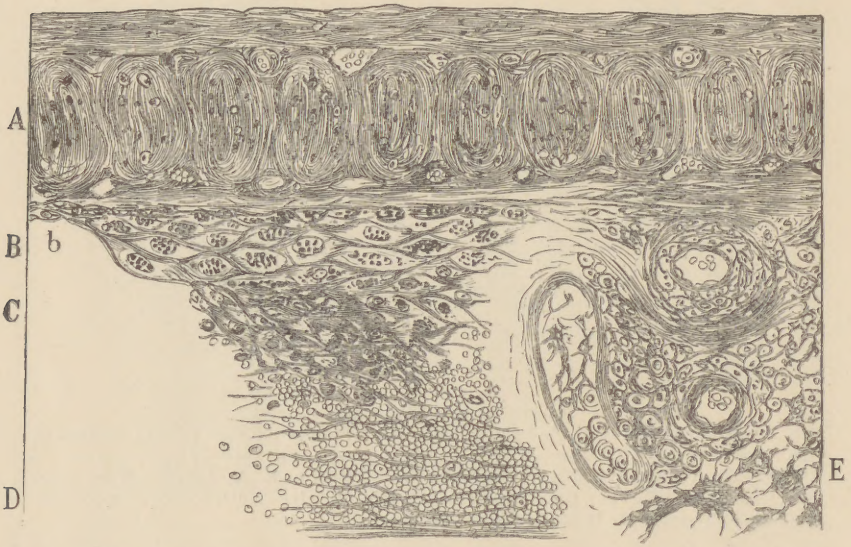
Endothelial and angiomatous beginning of the tumor. Whilst, as above stated, oval and net-celled forms are found, the chief interest centres in two genetic features encountered in the case. Since the literature on this subject is not accessible to me, and as I am wholly restricted for information to the text-books (Ziegler, Birch-Hirschfeld, Klebs), I do not know whether the facts which I am going to state have been previously observed or not.

At places quite distant from the seat of the tumor, in the dura mater, for instance, an inch from the margin of the remains of the tumor spoken of before, dural vessels are found, which show an enlargement of all of their elements. Arteries may be seen in which the endothelia and also the smooth muscle fibres are enormously enlarged. The same picture is often observed in the part of the brain which after the operation was invaded by the growth. The endothelium seems to grow more

actively, however, than the other constituents of the vessel, and soon the lumen is plugged with swelled-up endothelial cells, which in the course of time become flattened against each other.

Vaso-formative and hæmopoietic cells. Whilst thus the tumor spreads and invades the neighboring tissues by starting the endothelium of pre-existing vessels to proliferate, a different genetic factor may be observed in other parts. While examining the parts of the tumor not removed by the operation and the neighboring dura mater my attention was engaged by a vascular layer extending from the flattened tumor-remnants for a considerable distance underneath and along the dura mater.

FIG. 4.



A. Parietal (supra-arachnoidal) layer of the dura, showing a row of connective-tissue bundles cross cut. Vessels and lymph-spaces are seen along the upper and lower margins. B. Strata of pigmented cells, vaso-formative and hæmopoietic, starting from b, a piece of normal arachnoidea. C. Layer of capillary and blood transformation. D. Change to capillary angioma. E. Net-celled sarcoma from another portion of the dura. (HARTNACK, 3, vii.)

The chief interest centred in a layer of varying thickness of large spindle- or oval-shaped cells (Fig. 4) containing yellow granules. The cells gradually lose the yellow color, become larger, and on section represent a network with spindle-shaped spaces containing blood corpuscles. The spaces enlarge, and appear now as capillaries, the beginning of the sarcoma or angio-sarcoma. Those cells carrying pigmented granules seem to arise from the arachnoidea. I take them to be vaso-formative and hæmopoietic cells.

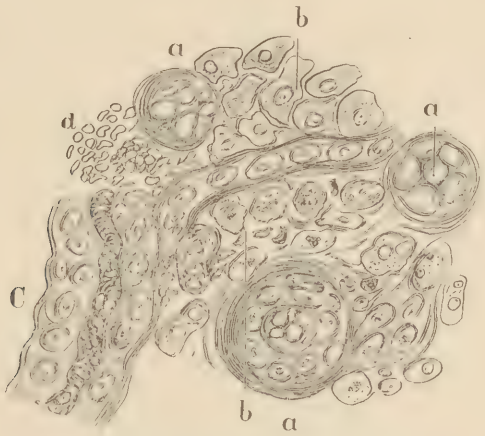
The blood, then, is formed *in loco*, together with the capillaries, a proposition which becomes at once a little more plausible and more easily

intelligible when we remember that the sarcomatous process is a counterfeit of embryonic connective-tissue formation, and that this process has been demonstrated by Ranvier in the tail of the tadpole.

In the recently-formed portions of the tumor vessels in the act of growing and new formation can be satisfactorily seen. This new formation is, of course, unlike that described for the angiomatous development from the dura and arachnoid. The walls of the vessels are thick, but transparent and frail. New vessels are formed by budding, and the new formation of blood-corpuscles seems to take place *pari passu* with that of the vessels. In these newly forming vessels, too, I have seen hemopoietic cells. They are not pigmented, however. The older hyaline vessels often resemble cactus, because of their clumsy branches. (See Fig. 3.)

The new formation of vessels and of blood in them is, however, not the only way of inaugurating the angiomatous process. In many parts of the dura I saw, besides the vascular proliferation, the filling up of lymph spaces with blood. Again, blood will take the place of solid connective tissue, melting, as it were, the intercellular substance, whereas the cells would appear constituting meshes in which the blood circulated.

FIG. 5.



Section through the invaded portion of the brain.

a, a, a. Vessels filling with proliferating endothelia. b, b. Enlarging and proliferating cells of the neuroglia. c. Artery whose muscular coat is undergoing the sarcomatous change. A capillary vessel to the right shows the same process. d. Extravasated red blood-corpuscles, some crescentic in shape; some are seen scattered among the neuroglia cells. (HARTNACK, 3, viii.)

The changes in the brain. There is an inflammatory and a neoplastic process demonstrable in the part of the brain nearest the former site of the tumor. I have no doubt that the injection of ammonia into the

arm during the operation and the subsequent suppuration had a great deal to do with the rapidly fatal termination of the case, although it must be admitted that the post-mortem findings were such as to preclude any possibility of complete recovery.

The surface of the brain was invaded by two kinds of micro-organisms —one a large coccus, the other a short, thick bacillus with rounded ends. No cultures were made, and a diagnosis is impossible.

Besides the inflammatory changes all the varieties of sarcomatous processes could be shown in even greater variety than in the membranes. Almost every field under the microscope reveals different pictures; the pathogenic scenery is constantly changing.

The changes occurring in the other organs—lungs and kidneys—were due to the infection caused by the hypodermatic needle.

Motor or sensori-motor area. The divergence of opinions as to the true functions of the central convolutions is as wide to-day as it was fifteen years ago. The most radical assertion on the subject has of late years been made by Flechsig and Hösel,[1] who claim that the so-called motor zone of the brain is at the same time the sensory centre (resp. reflex centre) of the posterior spinal columns, on the ground of embryologic and anatomo-pathologic findings. However, from the description given by these authors, it is evident that it is principally the *posterior* central convolution which is in anatomic relation with the sensory tracts of the spinal cord, an observation which fits in well with the clinical observations of neuro-surgery. These latter speak certainly a more emphatic language than embryologic researches of anatomic findings based on cases of long standing, especially if the lesions date back to childhood, as in the case of Flechsig and Hösel (porencephalic defect of fifty-two years' standing).

Our case flatly contradicts the theory of the sensori-motor character of the Rolandic region, in the sense that every motor area is at the same time the sensory cortical termination of the corresponding segment at the periphery of the body. It is, in its bearing on the question, well supplemented by a case published by Darkschewitsch,[2] who reports a monoplegia of the right arm due to circumscribed tubercular softening of a portion of the *centrum ovale* (left side) corresponding in the main to the middle third of the posterior central convolution. There were paralysis and greatly diminished sensibility of the right arm. No atrophy. The conclusion, then, to be drawn from the findings of our case and the one just mentioned would be that, so far as the middle third of the Rolandic region is concerned, the anterior portion is exclusively motor, the posterior chiefly sensory in function. Of the two, ours

[1] Die Centralwindungen, ein Centralorgan der Hinterstraenge. Neurol. Centrbl., 1890, p. 417.
[2] Zur Frage von den Sensibilitätsstörungen bei Herderkrankungen des Gehirns. Neurol. Centrbl., 1890, p. 714.

is the more telling and pronounced, owing to the exclusive involvement of the motor function. The diagram drawn by Darkschewitsch shows an encroachment of the lesion on a portion of the anterior third of the central at about the level of the tumor in our case. The question would at once become much clearer if an exclusively sensory disturbance due to cortical lesion could be demonstrated. This, to my knowledge, has not been done so far.

Outspoken sensory disturbances were also absent in a case of a large tumor to be detailed later on, reported by Pel, and one published by Dr. Mudd.[1] In the neurologic report, which I furnished for this case (echinococcus the size of a large hen's egg growing in the Rolandic region), I forgot to mention the fact that sensibility was intact, as ascertained by Dr. Fry and myself.

Import of the localizing symptoms. The clinical report says that the first manifestation of the developing disease consisted in the sudden appearance of weakness in the right shoulder and arm, the grip of the hand and the movements of the fingers being unimpaired. This monoparesis of the shoulder and arm was not transitory, but lasting. It was, therefore, of the highest possible localizing value.

According to Putnam,[2] monoplegias and paresis are of greater localizing value than monospasms and localized convulsions. The shoulder was first paralyzed. The probability, then, is that the starting point of the tumor was anteriorly to the anterior central convolution at the junction of the foot of the first and that of the second frontal convolution (shoulder centre), and that from this point it grew and spread back-up-downward. In the sequence, as the arm, wrist, and finger centres were pressed upon, the paralysis moved from joint to joint, a striking illustration of the distinctness of the various cortical areas controlling distinct sets of muscles which are wont to work together in harmony toward executing a certain movement. The spastic seizures, however, took place in the reverse sequence, beginning in parts last attacked, viz., the fingers. This is also in keeping with our knowledge about irritative lesions and their manifestations. In the light of the clinical history, as well as considering the actual state of the patient, no importance could be attached to the fact that the spasms began in the fingers. In view of the overtowering paralysis of the shoulder, the initial spasm of the fingers was of no localizing value, and could not be looked upon as a signal symptom from an operative point of view.

Absence of optic neuritis. The percentage of the cases of intra-cranial tumors in which double optic neuritis occurs has been variously estimated. That a case of suspected brain tumor becomes at once doubtful

[1] American Journal of the Medical Sciences, 1892, vol. ciii. p. 412.
[2] Intra-cranial Lesions. Transactions of the New York Medical Association, viii.

whenever this symptom, the most unequivocal and best known of intra-cranial growths, is found to be wanting, is almost an axiom in brain pathology. The strongly localizing symptoms, however, in our case could leave no doubt as to the existence of an intra-cranial tumor belonging to the minority, in which papillitis is wanting.

This is not the place to discuss the causes which lead to the production or prevention of optic neuritis in brain tumor; but it may not be amiss to point out the fact, exemplified once more in our case, that of all brain tumors not accompanied with papillitis, those of the large hemisphere and especially those occurring in the anterior portion of them, stand as to numbers at the head of the list.

Probably, also, the fact that the growth was not a brain tumor proper, and that, therefore, there was no irritating substance generated, which by some (Leber and Deutschmann), in addition to increased intra-cranial pressure, are held responsible for the production of papillitis, may partly account for the absence of this complication. Bramwell[1] remarks that in most of the cases of intra-cranial tumors in which optic neuritis is absent there is little or no evidence of increased pressure. This certainly applies fully to the case under discussion. There was neither hydrops internus nor distention of the sheaths of the optic nerves.

Absence of other classical symptoms of brain tumors. Besides optic neuritis, the strongest link in the chain of evidence establishing the diagnosis of cerebral tumor, the other characteristic symptoms were absent. There was no headache, vertigo, nausea, or vomiting, nor was there any mental hebetude or change of character.

The absence of headache in case of an intra-cranial growth of almost the size of a hen's egg may seem strange at first sight; but it becomes more plausible when it is remembered that there were no symptoms of any increase of intra-cranial pressure, notably no optic neuritis. For the same reason nausea and vomiting failed to set in, whilst vertigo is quite frequently absent in cases where the frontal region of a hemisphere is the seat of the lesion. All these symptoms were also absent in a large echinococcus cyst, reported by Mudd, and in Pel's case, referred to at another place. The spreading of the spasm to the face and mouth during the last attack of Jacksonian epilepsy showed how near Broca's convolution the disturbance had come. In spite of the relative remoteness of the tumor from this region, aphasic symptoms, by way of distant effect, would probably have manifested themselves had it not been for the fact that the patient was left-handed, and that therefore his speech-centres were on the right side of the brain. For the same reason there was no mental change, which, considering the size of the tumor-mass, would in all probability have shown itself if the patient had not been right-brained.

[1] Intra-cranial Tumors, p. 58.

Mechanism of the production of the destroying and irritating symptoms. The paralysis of the shoulder was the first symptom to set in; convulsions followed later on. This seems to be in keeping with the nature of the existing pathologic process. The pressure from the gradually-enlarging tumor caused probably at first a wasting of the underlying gray and, possibly, white substances, due principally to persistent anæmia. The discharging lesion which supervened in the course of time was probably the result of hemorrhages into the neighboring cortical and subcortical tissues.

Practical localization. Owing to the fact that a mono-paresis of the shoulder was the first symptom to appear, the inference is, that the starting-point of the dural tumor was over the shoulder-centre. Since, however, at the time the patient presented himself, the whole arm was paralyzed and involved in the spasm, the region of the skull corresponding to the middle third of the Rolandic region was selected as the site for operative interference. This anticipation was justified by the result. However, the beginning of the growth was probably at the shoulder-centre—*i. e.*, the foot of the second frontal convolution.

FIG. 6.

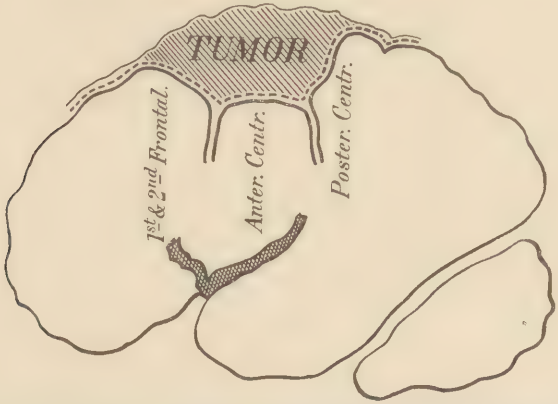


Diagram showing the relation of the tumor to the convolutions before operation. The dotted line represents the pia, the other, surrounding the tumor, represents the dura.

Statistics of cerebral muscular atrophy. There was prominent atrophy of the extremities of the left side. The muscles of the shoulder, notably the deltoid, supra- and infra-spinatus, were wasted. Muscular atrophy due to cerebral disease was first described by Todd.[1] Since then about twenty-one cases (most of them in late years, the present one included) have been registered and described, a remarkably small number considering the great prevalence of cerebral paralysis and the undoubted connection between muscular atrophy and brain lesion.

[1] Clinical Lectures on Paralysis, 1854.

Among the eighteen cases collected by Steiner[1] fourteen were due to causes other than neoplasms (hemorrhage, softening, inflammation, abscess), six were cerebral tumors, two, the last ones reported (Pel's and ours), were extra-cerebral, meaning meningeal growths.

Literature on cerebral amyotrophy due to brain tumors. Among the cases of cerebral muscle atrophy due to brain tumors I find the following mentioned in the literature: 1. Roth and Muratow,[2] glio-sarcoma gyri centr. dextr.; left arm atrophied. 2. Patella,[3] glioma corticis dextr.; left arm, small hand muscles, and left leg atrophied. 3. Quincke,[4] glioma gyri centr. dextr. utriusque. 4 Borgherini,[5] metastatic sarcoma gyri centr. sinistr. 5. Pel,[6] fibroma of meninges, over gyri centr. sinistr.

Our case would make the sixth in the literature on this subject. It may be that I have overlooked some cases, but those in which authors have laid stress on this unusual symptom are, I think, all included in this list. Probably lesser degrees of atrophy have been overlooked in similar cases. I believe that, by paying closer attention to this feature, a greater number of such cases would be recorded.

Muscular atrophy due to extra-cerebral lesion. The chief interest of our case, so far as the muscular atrophy is concerned, centres in the fact that the exciting cause was, strictly speaking, extra-cerebral; that it was a growth of the meninges and that by mere pressure, and probably wasting, of the cortex atrophy and electric change of reaction, to be spoken of later on, were brought on. There is only one case extant, published recently, in which features analogous to those observed in our case are described. Pel[7] details the history of a case in which, as in ours, the most striking, most important, and most constant symptoms of brain tumor (headache, nausea, vomiting, vertigo, optic neuritis) were absent although the brain mantle was compressed by a tumor the size of an egg. The paresis was on the right side. There was mental impairment. (It is not stated whether patient was right- or left-handed.) There was Jacksonian epilepsy, slight atrophy of the muscles of arm and leg. Complete intactness of sensibility and muscle-sense. Electric reaction normal. The tumor, about the form of a chestnut (probably horse-chestnut, as in our instance), was found over the arm-centre, flattening but not invading the convolutions beneath. It was a fibroma traversed by large lymph spaces growing from the pia mater.

Another remarkable case of early muscular atrophy, not mentioned

[1] Aus. de patholog. Inst. in Heidelberg. Deutsche Zeitschr. f. Nervenheilkunde, 1893, iii., 4. u. 5.

[2] Archives de Neurol., xxl. p. 296.

[3] Delle atrofie musculari secondarie, Padova, 1886.

[4] Deutsch. Arch. f. klin. Med., 1889, vol. xxxv. p. 371.

[5] Borgherini: Ueber einen Fall von Muskelatrophie, Neurolog. Centralbl., 1890, p. 545.

[6] P. K. Pel: Berl. klin. Wochenschr., 1894, No. 5.

[7] Loc. cit.

by Steiner, also due to extra-cerebral lesion, is that of a traumatic injury of the skull (fissure) corresponding to the anterior half of the upper parietal lobule, reported by J. R. Logan.[1] There was, besides a paralysis of the corresponding foot and leg (exclusive of the hip), a marked atrophy of the muscles of the calf.

The question of cortico-trophic centres and their location. In all of the cases of cerebral amyotrophy reported the arm was implicated, in some instances singly (in six), in the remainder other muscles participated. In none was there an absence of the arm paralysis when it was present in other parts. The only exception was the case of Logan, which, however is not entirely unquestionable as to precise localizing value (traumatic injury). It would be premature to deduce from this fact the conclusion that somewhere near the arm-centre there is a trophic cortical centre, possibly for all the muscle groups of the opposite side. The only deduction permissible is that, as the arm is generally most affected in cases of cerebral paralysis, so muscular atrophy of cerebral origin is most pronounced in the arm, while the face and the leg may remain intact. The fact that there was absolute absence of degenerative changes of the cortico-muscular tract and the ganglion cells of the anterior horn would point to direct trophic fibres passing from some portion of the brain to the muscles. These centres, however, are, so far, only hypothetical. At all events, it is rather premature to conclude, as Luzzato (remark made by Steiner, *loc. cit.*) has done, that early atrophy in hemiplegias means involvement of the cortex.

There is at least one case in neurologica literature which points to quite a different direction.[2]

This case would tend to prove, as the title indicates, that the gyrus fornicatus is not only the centre of sensation in accordance with the experiments of Horsley and Shaefer, but also of nutrition.

This observation, however, stands so isolated that it cannot count against the mass of evidence pointing to the Rolandic region as the probable seat of the musculo-trophic centres. Still, this is as yet an obscure subject.

Amyotrophy without the customary spinal lesions. What is particularly remarkable is the fact that there was no trace of a descending degeneration of the pyramidal tracts, nor were the ganglionic cells of the anterior horn altered to any considerable extent, contrary to Charcot's teaching, who holds disease or atrophy of these cells responsible for muscular atrophy occurring in cerebral paresis. Similar cases have

[1] Fracture of the Skull with Localized Paralysis. Lancet, September 29, 1883.

[2] Saville: On a Case of Anæsthesia and Trophic Changes Consequent on a Lesion Limited to the Gyrus Fornicatus and Part of the Marginal Convolution. Brain, Summer and Autumn, 1891.

been observed by Quincke,[1] who established the absence of descending degeneration in the corresponding pyramidal tracts and the integrity of the large ganglionic cells of the anterior in two cases. Our case furnishes an analogy to those of Quincke. One cannot help thinking of a "dynamic" influence of the cortical cells, however distasteful such a qualification may appear to the naturalist and anatomist.

Comparative frequency of infantile and adult cerebral amyotrophy. It is a notable fact that cerebral amyotrophies in children are very frequent, as compared with the number met with in adults. Steiner[2] suggests that in children there is a certain dependence of the ganglion cells of the anterior horns on the ganglion cells of the brain; hence, cerebral atrophies are not rare. In adults the spinal ganglion cells are more independent, hence the rarity of muscular atrophies in the cerebral palsies of adults. If they do occur, they find their analogy, according to Steiner, in the infantile type. This, of course, is not an explanation, but a suggestion; besides, the correctness of the statement may be questioned. For, functionally, at any rate, the spinal cord is the more independent of the brain the younger the animal, and *vice versa*. For what is true phylogenetically, that the lower the order of the vertebrates the closer the approach to the spinal-cord animal, down to the lancet fish, which has almost no brain, is also true ontogenetically. The closer the approach to adult life of a vertebrate, the greater the subordination of the cord to the brain.

Post operative sensory disturbances. In this case, as in several others of the same type I had an opportunity of observing, impairment of sensation set in immediately after the operation, whilst no disturbance of this kind could be made out before. Here was a tumor of considerable size pressing on the whole of the middle third of the Rolandic area, producing complete paralysis of the corresponding arm and shoulder, and yet there was not the slightest trace of deficient sensation. But immediately following the operation there was marked anæsthesia. Such observations are in the highest degree perplexing, proving and disproving in the same individual the sensory qualities of the Rolandic region.[3]

Electrical reaction of the nerves and muscles. It is a well-known fact that in cerebral paralysis electric irritability of the affected muscles is not essentially altered from the normal types; that, if any change exists at all, it occurs in cases of very long standing, and consists in a simple lowering of muscular or neuro-muscular irritability corresponding to

[1] Ueber Muskelatrophie nei Gehirnerkrankungen. Deutsches Arch. f. klin. Med., 1888, Bd. xlii., quoted by Eisenlohr. Muskelatrophie und electrische Erregbarkeitsveränderungen bei Hirnherden. Neurol. Centralbl., 1890, p. 1.

[2] Loc. cit.

[3] Cf. Bremer and Carson: A Case of Brain Tumor, etc. THE AMERICAN JOURNAL OF THE MEDICAL SCIENCES, September, 1890, vol. c. p. 240.

the intensity of the atrophy. In this case, electric exploration of the nerves and muscles of the affected side yielded in brief the following results: There was a slight lowering of the electric irritability, both faradic and galvanic. Galvanic reaction of the right ulnar nerve was changed, A C C > C C C. Of course, this is simply a reversion of the type, and degenerative reaction could not be diagnosticated on this finding, the characteristic sign of this being the slow, lazy contraction. This I could not establish in any of the muscles.

Only in a few cases of cerebral muscular atrophy have the muscles and nerves been electrically tested. So far such tests are chiefly of theoretic value. They are the more so ever since the electro-diagnostic differentiation between central and peripheral paralysis has been demonstrated to stand on a weak foundation. The establishment of degenerative reaction, even in hysterical muscular atrophy has almost dealt the death-blow to the formerly highly-prized diagnostic value of the reaction of degeneration.

Causation. Whether there was any causal relation between the fall on the head and the development of the tumor later on, can be only a matter of surmise. But the often-repeated observation, that brain tumors, principally gliomatous, but not infrequently also sarcomatous, have followed direct and indirect injuries of the head, point to the accident as a probable causative element.

Remarks. The development and progress of the endotheliomatous process in the brain after the operation must have been a very rapid one. As remarked before, the brain-substance pressed upon by the tumor seemed intact at the time of the operation. Manipulation incident to the removal gave, no doubt, the impetus to the more rapid spreading of the morbid growth in the meninges and to the invasion of the brain-substance proper, which before the operation must have been free from tumor invasion. That in this case a good chance of complete success was missed, there can be no manner of doubt. If, instead of frittering away valuable time with the superfluous galvanic and worse than useless faradic current, and the bromide treatment—if, instead of all this, the localizing value of the paresis of the shoulder had been properly appreciated, the tumor might have been removed at a time when its dimensions admitted of a liberal excision of healthy dura mater around it, and when the compression of the brain-substance was not too far advanced to preclude the possibility of a *restitutio ad integrum*. As it was, the filling of the cavity left by the removal of the tumor was probably, in spite of the tampon, so rapid that numerous capillary hemorrhages must have taken place in and around the compressed area of the brain. The softening of nerve tissue was in part due to these hemorrhages.

The scientific importance of our case is somewhat impaired by the

patient surviving the operation for over two weeks. During that time inflammatory changes and invasion of the malignant growth into the brain very much obscured and complicated the anatomo-pathologic conditions giving rise to the symptoms during life. It would, above all, have been of importance to ascertain to what degree and in what manner the cortex was changed. The topography of these changes might possibly have thrown more light on the question of a trophic cerebral centre.